8 to 14. The post-mortem findings in two cases are described. Two deaths occurred in previously healthy children and three in children with a pre-existing illness.

We wish to thank Dr. E. L. Simons for the pathological and post-mortem reports.

REFERENCES

Atkinson, E. C. (1957). *Brit. med. J.*, 2, 821.
Cantcr. J. (1957) Ibid., 2, 1112.
Fielding, G (1957) Ibid., 2, 763.
Mann, R., and Pasha, M. A. (1957). Ibid., 2, 939.
Vital Statistics (1957). Ibid., 2, 1056.

A WOMAN WITH THE STIFF-MAN SYNDROME*

BY

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"A curious condition of progressive, fluctuating muscular rigidity and spasm has recently been described by F. P. Moersch and H. W. Woltman, of the Mayo Clinic, as the 'stiff-man' syndrome." These were the opening words of an annotation in the *British Medical Journal*† that I was reading in January, 1957, when I realized they were a clear description of the case of a woman whose extraordinary spasms had puzzled me ten years previously. Luckily my ward sister remembered her name; the notes were traced and revealed that the lady had continued to suffer from "hysterical spasm" which had disabled and deformed her till she had become a bedridden cripple cared for in Leamington Park—a geriatric annexe to the Central Middlesex Hospital. Dr. A. Abdullah, the physician in charge, kindly allowed me to review her case. Everything fitted the description in the annotation, and later the annotator himself came and agreed that this woman had the stiff-man syndrome.

I wish to report this case (*a*) because I made the diagnosis ten years late; (*b*) because it appears to be the first case of stiff-man syndrome reported in England; (*c*) because it shows that syndromes are sometimes labelled hysterical for no better reason than that they are not yet officially described, even in this case where the patient's hysterical spasms were so titanic (and that is not a misprint for tetanic) that with their power she bent a Smith-Petersen pin and fractured a steel plate and screws; (*d*) because it shows the value of annotations in keeping physicians in touch with rarities and novelties: and (*e*) because since I recognized the diagnosis the patient has died and the necropsy findings are now known.

Case History

A housewife of 50 was admitted to hospital in May, 1946, with painful spasms in her legs, abdomen, and back which had been present for two days. She gave a history of "lumbar fibrositis" for six years, treated at another hospital with physiotherapy, and for three years had had increasing back stiffness, which confined her to the house, punctuated by acute attacks of "cramps" in the abdomen, legs, and back similar to the one which brought her in.

Examination showed a dull-witted, distraught woman who groaned continually. The muscles of her abdomen, thighs, and calves were rigid and hard. Her whole body was so stiff she could scarcely move. At intervals acute spasms would come on her with tetanic severity. She would scream and sweat, clutching bystanders, contorting her limbs, and becoming pulseless for a while.

Apart from a generalized rigidity and these remarkable muscular spasms there were no findings except for a colostomy which had been done for ulcerative colitis eight years previously. She had had no bowel troubles since.

She stayed in hospital a month. Every relevant (and irrelevant) biochemical and radiological investigation was negative. Her spasms gradually became less and she was discharged on June 11. I wrote to her doctor, "I am unable to make a definite diagnosis in this case." I made an indefinite diagnosis of spasms secondary to spinal arthritis with hysterical overlay.

Two years later she was admitted to a mental hospital with depression and "troublesomeness." The latter was shown by attacks of spasm and screaming. A month after admission she felt her femur snap during a spasm. The *x*-ray film showed a subcapital fracture of the neck and she was transferred back to the Central Middlesex Hospital, where a 3½-in. (9-cm.) Smith-Petersen pin was inserted. The notes record that next day she was "very troublesome with emotional crises" and subsequent *x*-ray films showed gross varus deformity with bending of the pin and a pertrochanteric fracture through the pin track.

The pin was removed and a mail plate inserted. Again her troublesomeness continued, the deformity recurred, and the screw and plate were broken. A third operation was done two weeks later, but gradually the gross adduction deformity of the right hip recurred. Many attempts were made to mobilize her, but they provoked further spasms and screaming. She developed increasing varus deformity of the feet and her rigidity became worse than ever. She became a chronic case and stayed for five years in the wards before being discharged to a geriatric annexe.

In May, 1955, she was readmitted to the Central Middlesex Hospital for assessment. The houseman's note was: "Condition worsening. Hysterical outbursts on little or no provocation—hyperpnoea, rolling up eyes, clutching fiercely at bed, locker, or bystanders, voluntary rigidity of muscles with opisthotonos, becoming worse if soothed, gradually quietening if ignored."

I was asked to see her, and wrote: "I am most interested to see this woman after nearly ten years. Her condition seems very much the same as when I last saw her. I still feel there must be an organic basis for her symptoms—a sort of dystonia musculorum deformans but without movement."

A psychiatrist was later called, and wrote: "Whatever her difficulties are she appears to be completely lacking in insight and therefore psychologically inaccessible. It is obvious that her rehabilitation is a formidable proposition; she appears to have satisfied some emotional need in her illness. I do not think that any psychiatric approach is likely to be effective and that her treatment is mainly one of general management. I feel the prognosis is bad."

The deformity of the feet induced by constant spasm had now produced a gross talipes equinovarus. This was partially corrected by manipulation and plaster, and while in plaster she succeeded in walking a little with a stick, but when the plaster was removed the deformity recurred, so in November, 1955, an arthrodesis of the left ankle was done and further attempts to mobilize her were made. These achieved little success, so she was transferred to Leamington Park, where they continued for another six months to try to get her up. All efforts at salvage were abandoned finally in August, 1956, when the house-physician wrote: "Very difficult and uncooperative. Prefers to stay in bed. As this makes it much easier for sister and as the patient obviously does not want to get better, it is time we realized we have lost this battle."

Thereafter she remained bedridden—rigid and deformed but moderately content, provided she was not moved, when "hysterical" spasm would recur.

Four months after therapeutic defeat had been openly admitted I visited her again because I had read the annotation mentioned above. The many tattered pages of notes

*I cannot avoid the paradoxical title; those who named this illness called it "stiff-man syndrome"—stiff-person syndrome might be a better name.

†*Brit. med. J.*, 1956, 2, 1478.

were re-read and condensed and physical examination was repeated. The story outlined here was typical of the syndrome described by Moersch and Woltman, and physical examination was as unrevealing as it had been in their cases.

All there was to find was a hypertrophy of the muscles of the neck and trunk with board-like rigidity of these muscles, the abdominal muscles, and the muscles of the lower limbs. The legs were contorted into grossly misshapen deformities, one ankle had varus of such degree that the sole of the foot was facing upwards.

The urine occasionally revealed a trace of glucose which 4 out of Moersch and Woltman's 14 cases had shown.

To be able to identify and name the condition gave the patient no benefit whatever. No doubt, had she known Matthew Arnold's lines she might have expressed the same wish as he did:

> Nor bring to watch me cease to live
> Some doctor, full of phrase and fame,
> To shake his sapient head and give
> The ill he cannot cure—a name.

With Dr. Abdullah's kind permission the patient was brought over to one of this hospital's weekly clinical meetings at the beginning of January, 1957. When I exposed her body and gently moved one limb to show the muscular rigidity this provoked such dramatic and widespread spasm that she was clearly in great agony, crying out loudly, gripping bystanders till her nails dug into them, and sweating visibly and profusely. It appeared that the one-mile ambulance journey had brought her to a climax of acute spasm, and the nurses who accompanied her begged that she might be given morphine for the journey back. We at the meeting felt guilty that a clinical demonstration had unintentionally produced so much suffering. She returned to Leamington Park under heavy sedation and recovered in a few days. In April a course of "tolseram," 1 g. t.d.s. gradually increasing to 3 g. t.d.s., was tried for one month. It made no difference. Thereafter she remained in her usual rigid bedfast condition for another six months, till her unexpected death on June 18. The house-physician's note makes a suitable epitaph: "Commenced 'hysterical turn' at 10.30 p.m. on June 17, 1957, and settled down at 2 a.m. Found dead at 3 a.m." Her hysteria had at least been awarded the posthumous dignity of inverted commas.

Post-mortem Findings

A complete microscopical examination showed no changes except those attributable to age and atherosclerosis. The only significant changes were in the muscles. These changes, which showed best in the sacrospinalis muscle and hardly at all in the pectoral muscles, were: (*a*) Abnormal variations in the size and staining reaction of the muscle fibres; the swollen fibres were pale and lacking in striation. (*b*) Increase of the connective tissues both collagen and elastic fibres, principally the former. (*c*) Localized areas of atrophy and fibrosis of muscle accompanied by nuclear aggregates and lymphorrhages.

Dr. C. Treip reported that these muscular changes appeared to be primary and not related to any change in the central nervous system. This supports the idea that this illness may be a type of myopathy. Nevertheless it is possible the changes are secondary to spasm or to prolonged bed rest, and the true pathology still remains uncertain.

Comment

The way I have described this case is a little unorthodox, but I thought it would make it easier to read. Reports of single cases of rare diseases are not usually exciting; this was an exciting and rather fearful case, so I have tried deliberately to describe it in a way that might convey this.

Summary

A woman of 50 suffered from progressive, fluctuating muscular rigidity and spasms for over 15 years. The spasms caused a fractured leg and broke the steel fittings used to repair it. For many years the spasms were labelled hysterical. Six months before death a diagnosis of stiff-man syndrome was made because of the clear resemblance to Moersch and Woltman's cases. No treatment did any good. She died after a severe spasm, and post-mortem examination showed muscular changes, which suggests this illness is some form of myopathy.

I am indebted to Dr. A. Abdullah for his courtesy in allowing me to follow up this patient in his wards after she left my care; to my registrar, Dr. L. E. Lennard-Jones, for his general assistance; and to Dr. C. Treip for the post-mortem report and the extensive microscopy studies.

Medical Memoranda

Two Cases of Bullous Chicken-pox

Though bulla formation is mentioned as an occasional complication of chicken-pox, it is rarely seen. These cases are interesting in that one aroused a transient fear of phenol poisoning and that both made a complete recovery without residual scarring, in spite of the alarming picture which each presented when the disease was at its height.

On August 13, 1956, in a small housing estate in Surrey, where chicken-pox was epidemic, a girl aged 8 years, successfully vaccinated in infancy and with no history of chicken-pox or herpes, was seen at the commencement of what appeared to be a mild attack of chicken-pox. There were about 20 pocks on the trunk and a dozen on the face, scalp, and neck; none on any mucous membranes. Treatment was started with aspirin and local application of 1% phenol in calamine lotion.

When the patient was next seen, three days later (fourth day of illness), there were several large discrete bullae, each overlying the site of a pock, on brow, nuchal region, left axilla, left elbow, upper abdomen, and vulva. These were filled with a turbid fluid, and varied in area from 7 by 5 cm. to 3 by 2 cm. Each was surrounded by a zone of erythema 1 to 2 cm. wide, and the appearance was of a number of localized burns, as from a hot-water bottle. Scattered over the intervening skin were normal pocks in an unexpectedly advanced state of involution.

Though it seemed unlikely, when such a small proportion of treated pocks had proceeded to bulla formation, the possibility was considered that the patient was abnormally sensitive to the low percentage of phenol in the lotion applied or that the percentage had been accidentally increased in the course of dispensing. Local dressing with 0.5% cetrimide cream was substituted and tetracycline administered orally in the hope of reducing septic complications.

The suspicion of phenolic poisoning was renewed on the following day (fifth day of illness), when there was oedema of the eyelids and face sufficient to close the left eye; but there was no oliguria, and the urine was free from protein and normal on microscopy, so the oedema was assumed to have arisen by drainage from tissues surrounding the large bulla in the left frontal region. There was similar oedema of the vulva. By now nearly all the bullae had ruptured and a central pock could be seen in each of the raw areas revealed.

On the seventh day of illness improvement was apparent. Oedema was lessened and the denuded areas were healing without obvious secondary infection. By the end of the second week the patient was ambulant, and at four weeks there remained only two small cicatrices such as may frequently be found after an uncomplicated attack.

On August 26, 13 days after the onset in the first patient, her brother, aged 17 months, developed bullae on both upper eyelids, scalp, thighs, and one finger, with a heavier crop of normal pocks on the trunk than his sister had had. There had been no contact with the first patient from the